

| National Imaging Associates, Inc.*   |  |
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| Clinical guideline<br>CT HEART<br>CT HEART Congenital<br>(Not including coronary arteries) | Original Date: September 1997                                  |
| CPT Codes: 75572, 75573  | Last Revised Date: <del>March-February 2022</del> <sup>1</sup> |
| Guideline Number: NIA_CG_025   | Implementation Date: January 2023 <sup>2</sup>                 |

## GENERAL INFORMATION

It is an expectation that all patients receive care/services from a licensed clinician. All appropriate supporting documentation, including recent pertinent office visit notes, laboratory data, and results of any special testing must be provided. All prior relevant imaging results and the reason that alternative imaging cannot be performed must be included in the documentation submitted.

## INDICATIONS FOR HEART COMPUTED TOMOGRAPHY (CT)<sup>1, 2</sup>

~~(Douglas, 2011; Taylor, 2010)~~

### Congenital Heart Disease<sup>3</sup>

~~(Sachdeva, 2020)~~

For all indications below, either CT or CMR can be performed:

- All congenital lesions: prior to planned repair and for change in clinical status and/or new concerning signs or symptoms
- Patent Ductus Arteriosus: routine surveillance (1-2 years) in a patient with postprocedural aortic obstruction
- Aortic Stenosis or Regurgitation: routine surveillance (6-12 months) in a child with aortic sinus and/or ascending aortic dilation with increasing size
- Aortic Coarctation and Interrupted Aortic Arch:
  - Routine surveillance (3–5 years) in a child or adult with mild aortic coarctation
  - Post procedure (surgical or catheter-based) routine surveillance (3–5 years) in an asymptomatic patient to evaluate for aortic arch aneurysms, in-stent stenosis, stent fracture, or endoleak
- Tetralogy of Fallot:

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- Routine surveillance (2–3 years) in a patient with valvular or ventricular dysfunction, right ventricular outflow tract obstruction, branch pulmonary artery stenosis, arrhythmias, or presence of an RV-to-PA conduit
- D-Loop Transposition of the Great Arteries (postoperative):
  - Routine surveillance (3–5 years) in an asymptomatic patient
  - Routine surveillance (1–2 years) in a patient with dilated aortic root with increasing size, or aortic regurgitation
  - Routine surveillance (3–12 months) in a patient with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, LVOT obstruction, or arrhythmias
- Congenitally Corrected Transposition of the Great Arteries:
  - Unrepaired: routine surveillance (3–5 years) in an asymptomatic patient
  - Postoperative: routine surveillance (3–5 years) in an asymptomatic patient
  - Postoperative anatomic repair: routine surveillance (6–12 months) in a patient with valvular or ventricular dysfunction, right or left ventricular outflow tract obstruction, or presence of an RV-to-PA conduit
  - Postoperative physiological repair with VSD closure and/or LV-to-PA conduit: routine surveillance (3–12 months) in a patient with ≥moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit dysfunction
- Truncus Arteriosus: routine surveillance (1–2 years) in an asymptomatic child or adult with ≥ moderate truncal stenosis and/or regurgitation
- Single-Ventricle Heart Disease (includes hypoplastic left heart syndrome, double-inlet LV, double-inlet RV, mitral atresia, tricuspid atresia, unbalanced A-V septal defect):  
postoperative routine surveillance (13–25 years) in an asymptomatic patient:  
~~postoperative routine surveillance (3–5 years) in an asymptomatic patient~~
- ~~Hypoplastic Left Heart Syndrome: prior to planned palliative procedures and for post-operative routine surveillance (2–3 years) following Fontan procedure).~~
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## Cardiomyopathy

- Quantification of myocardial (muscle) mass (CMR or CT)
- Assessment of right ventricular morphology in suspected arrhythmogenic right ventricular cardiomyopathy, based upon other findings such as:
  - Nonsustained VT
  - Unexplained syncope
  - ECG abnormalities
  - First-degree relative with positive genotype of ARVC  
(either, but CMR is superior to CT)<sup>4, 5</sup> ~~(Marcus, 2010; te Riele, 2015)~~

## Valvular Heart Disease

- Characterization of native or prosthetic valves with clinical signs or symptoms suggesting valve dysfunction, when TTE, TEE, and/or fluoroscopy have been inadequate<sup>6</sup> ~~(Doherty, 2017)~~

- Evaluation of RV function in severe TR, including systolic and diastolic volumes, when TTE images are inadequate and CMR is not readily available
- Pulmonary hypertension in the absence of severe valvular disease
- Evaluation of suspected infective endocarditis with moderate to high pretest probability (i.e., staph bacteremia, fungemia, prosthetic heart valve, or intracardiac device), when TTE and TEE have been inadequate
- Evaluation of suspected paravalvular infections when the anatomy cannot be clearly delineated by TTE and TEE<sup>7</sup> ~~(Nishimura, 2014)~~

### Evaluation of Intra- and Extra-cardiac Structures

- Evaluation of cardiac mass, suspected tumor or thrombus, or cardiac source of emboli, when imaging with TTE and TEE have been inadequate
- Re-evaluation of prior findings for interval change (i.e., reduction or resolution of atrial thrombus after anticoagulation), when a change in therapy is anticipated<sup>6-8</sup> ~~(Baumgartner, 2017; Doherty, 2017; Nishimura, 2014)~~
- Evaluation of pericardial anatomy, when TTE and/or TEE are inadequate or for better tissue characterization of a mass and detection of metastasis [CMR superior for physiologic assessment (constrictive versus restrictive) and tissue characterization, CT superior for calcium assessment]<sup>9, 10</sup> ~~(Klein, 2013; Pennell, 2010)~~

### Electrophysiologic Procedure Planning<sup>2</sup>

~~(Taylor, 2010)~~

- Evaluation of pulmonary venous anatomy prior to radiofrequency ablation of atrial fibrillation and for follow-up when needed for evaluation of pulmonary vein stenosis
- Non-invasive coronary vein mapping prior to placement of biventricular pacing leads

### Transcatheter Structural Intervention Planning

- Evaluation for transcatheter aortic valve replacement (TAVR)<sup>6, 11, 12</sup> ~~(Doherty, 2017; Otto, 2017; Schoenhagen, 2010)~~
- When TTE and TEE cannot provide adequate imaging, CT imaging can be used for planning: robotic mitral valve repair, atrial septal defect closure, left atrial appendage closure, ventricular septal defect closure, endovascular grafts, and percutaneous pulmonic valve implantation<sup>12, 13</sup> ~~(Pison, 2015; Schoenhagen, 2010)~~
- Evaluation for suitability of transcatheter mitral valve procedures, alone or in addition to TEE<sup>14</sup> ~~(Wunderlich, 2018)~~

### Aortic Pathology<sup>6-8, 15-20</sup>

~~(Baumgartner, 2014; Bhawe, 2018; Doherty, 2017, 2019; Erbel, 2014; Hendel, 2006; Hiratzka, 2010; Nishimura, 2014; Svensson, 2013)~~

- CT, MR, or echo can be used for screening and follow-up, with CT and MR preferred for imaging beyond the proximal ascending thoracic aorta in the following scenarios:
  - Evaluation of dilated aortic sinuses or ascending aorta identified by TTE
  - Suspected acute aortic pathology, such as dissection

- Re-evaluation of known aortic dilation or aortic dissection with a change in clinical status or cardiac examination or when findings would alter management
- Screening first-degree relatives of individuals with a history of thoracic aortic aneurysm or dissection, or an associated high-risk mutation for thoracic aneurysm in common
- Screening second-degree relative of a patient with thoracic aortic aneurysm, when the first-degree relative has aortic dilation, aneurysm, or dissection
- Six-month follow-up after initial finding of a dilated thoracic aorta, for assessment of rate of change
- Annual follow-up of enlarged thoracic aorta with size up to 4.4 cm
- Biannual (twice/yr) follow-up of enlarged aortic root  $\geq 4.5$  cm or showing growth rate  $\geq 0.5$  cm/year
- Patients with Marfan's syndrome may undergo annual imaging with CT, MRI or TTE, with increase to biannual (twice-yearly) when diameter  $\geq 4.5$  cm or when expansions is  $> 0.5$  cm/yr
- Patient with Turner's syndrome ~~patients~~ should undergo initial imaging with CT, MRI, or TTE for evidence of dilatation of the ascending thoracic aorta. If imaging is normal and there are no risk factors for aortic dissection, repeat imaging should be performed every 5 - 10 years, or if otherwise indicated. If the aorta is enlarged, appropriate follow-up imaging should be done according to size, as above
- Evaluation of the aorta in the setting of a known or suspected connective tissue disease or genetic condition that predisposes to aortic aneurysm or dissection (i.e., Loeys-Dietz, Ehlers-Danlos), with re-evaluation at 6 months for rate of expansion. Complete evaluation with CMR from the cerebrovascular circulation to the pelvis is recommended with Loeys-Dietz syndrome.

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## BACKGROUND

- Cardiac computed tomography (Heart CT) images the cardiac chambers, great vessels, valves, myocardium, and pericardium to assess cardiac structure and function, particularly when echocardiography (transthoracic echocardiography and transesophageal echocardiography) cannot provide adequate information
- CT imaging can be used for assessment of:
  - Structures of the heart (e.g., chambers, valves, great vessels, masses, ~~etc.~~), as in this guideline
  - Quantitative level of calcium in the walls of the coronary arteries, in the separate coronary artery calcium (CAC) scoring guideline

## OVERVIEW<sup>2</sup>

~~(Taylor, 2010)~~

### Imaging in Congenital Heart Disease

Echocardiography is often utilized for initial assessment of congenital heart disease. However, if findings are unclear or need confirmation, CMR or CT can be useful ~~(Sachdeva, 2020)~~.<sup>3</sup>

### CT and Cardiac Masses

CT and CMR are used to evaluate cardiac masses, describing their size, density, tissue characteristics, and spatial relationship to adjacent structures.

### CT and Pericardial Disease

While echocardiography is most often used in the initial examination of pericardial disease, CT and CMR can evaluate pericardial thickening and masses which are often detected initially with echocardiography. CT and CMR can accurately define the site and extent of masses, e.g., cysts, hematomas, and neoplasms ~~(Klein, 2013)~~.<sup>9</sup>

### Abbreviations

|                    |   |
|--------------------|---|
| ARVD/C             | Arrhythmogenic right ventricular dysplasia/-cardiomyopathy                    |
| CABG               | Coronary artery bypass grafting surgery                                       |
| CAD                | Coronary artery disease   |
| CCS                | Coronary calcium score  |
| CCT                | Cardiac (heart) CT  |
| CHD                | Coronary heart disease  |
| CMR                | Cardiac magnetic resonance (imaging)  |
| CT                 | Computed tomography   |
| CTA                | Computed tomography angiography   |
| ECG                | Electrocardiogram   |
| EF                 | Ejection fraction   |
| HF                 | Heart failure   |
| <b><u>LVOT</u></b> | <b><u>Left ventricular outflow tract</u></b>                                  |
| MI                 | Myocardial infarction   |
| MPI                | Myocardial perfusion Imaging or cardiac nuclear imaging                       |
| MR(I)              | Magnetic resonance (imaging)  |
| <b><u>PA</u></b>   | <b><u>Pulmonary artery</u></b>  |
| PCI                | Percutaneous coronary intervention  |
| PVML               | Paravalvular mitral leak  |
| RV                 | Right ventricle   |
| SE                 | Stress <del>e</del> Echocardiogram  |
| TAVR               | Transcatheter <del>a</del> Aortic <del>v</del> Valve <del>r</del> Replacement |
| TMVR               | Transcatheter mitral valve replacement  |
| TR                 | Tricuspid regurgitation   |
| <b><u>TEE</u></b>  | <b><u>Transesophageal echocardiography</u></b>                                |
| TTE                | Transthoracic echocardiography  |
| <b><u>VT</u></b>   | <b><u>Ventricular tachycardia</u></b>   |

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## POLICY HISTORY

| Date                             | Summary  |
|----------------------------------|--|
| <u>February 2022<del>3</del></u> | <u>Listed clinical spectrum comprising single-ventricle heart disease to include: hypoplastic left heart syndrome, double-inlet LV, double-inlet RV, mitral atresia, tricuspid atresia, unbalanced A-V septal defect</u><br><del>Added indications to single-ventricle heart disease: hypoplastic left heart syndrome, double inlet LV, double inlet RV, mitral atresia, tricuspid atresia, unbalanced A-V septal defect</del><br><u>hypoplastic left heart syndrome</u>   |
| March 2021                       | No changes   |
| March 2020                       | <ul style="list-style-type: none"> <li>• Added general information section as Introduction which outlines requirements for documentation of pertinent office notes by a licensed clinician, and inclusion of laboratory testing and relevant imaging results for case review</li> <li>• Extensive update to the indications for Congenital Heart Disease to include the following: <ul style="list-style-type: none"> <li>○ For all indications noted, either CT or CMR can be done</li> <li>○ All lesions: evaluation prior to planned repair and evaluation for change in clinical status and/or new concerning signs or symptoms</li> <li>○ Specific indications based on lesion were added with interval and criteria for repeat imaging included</li> </ul> </li> <li>• Added separate section for infective endocarditis</li> <li>• Removed tables of aortic diameter norms and suggested follow-up imaging</li> <li>• Edits to background with removal of table outlining radiation exposure and comment</li> <li>• Edits to overview included, with removal of the following: <ul style="list-style-type: none"> <li>○ CT and CMR provide 3D anatomic relationship of the blood vessels and cardiac anatomic structures</li> <li>○ Discussion of cardiac myxoma</li> </ul> </li> <li>• Updated and added new references</li> </ul> |
| July 2019                        | <ul style="list-style-type: none"> <li>• Added the following indication: Evaluation of anomalous thoracic arteriovenous vessels, such as transposition of the great arteries, when magnetic resonance imaging (MRI) cannot be performed</li> <li>• For valvular heart disease added indication for pulmonary hypertension in the absence of severe valvular disease</li> <li>• Removed indication: to assess degree of calcification in calcific aortic stenosis</li> <li>• For evaluation of intra- and extra-cardiac structures, the following indication was added: Re-evaluation of prior findings for interval</li> </ul>   |

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|  | <p>change (i.e. reduction or resolution of atrial thrombus after anticoagulation), when a change in therapy is anticipated</p> <ul style="list-style-type: none"> <li>• Removed section: scenarios in which heart CT is not indicated</li> <li>• Removed statement: CT imaging is competitive with MRI, but left in table in comparing two modalities (removed cost comparison)</li> </ul> |
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**Reviewed / Approved by NIA Clinical Guideline Committee**

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Reviewed / Approved by NIA Clinical Guideline Committee



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